

What Is Claimed:

1           1. A method for treating a subject suffering  
2 from phenylketonuria and/or phenylalanemia, said method  
3 comprising:

4           enterally administering to the subject a LNAA  
5 supplement in which the weight ratio of Leu to Val is  
6 greater than 2:1.

1           2. A method according to claim 1, wherein the  
2 LNAA supplement is substantially free from phenylalanine.

1           3. A method for treating a subject suffering  
2 from phenylketonuria and/or phenylalanemia, said method  
3 comprising:

4           enterally administering to the subject a LNAA  
5 supplement in which the weight ratio of Leu to iLeu is  
6 greater than 3:1.

1           4. A method according to claim 3, wherein the  
2 weight ratio of Leu to Val in the LNAA supplement is  
3 greater than 2:1.

1           5. A method according to claim 4, wherein the  
2 LNAA supplement is substantially free from phenylalanine.

1           6. A method for treating a subject suffering  
2 from phenylketonuria and/or phenylalanemia, said method  
3 comprising:

4           enterally administering to the subject a LNAA  
5 supplement which comprises one or more LNAAs and which  
6 further comprises Lys.

1               7. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu.

1               8. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu and wherein the weight  
3 ratio of Leu to iLeu in the LNAA supplement is greater  
4 than about 0.5:1.

1               9. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu and wherein the weight  
3 ratio of Leu to iLeu in the LNAA supplement is greater  
4 than 3:1.

1               10. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu and wherein the weight  
3 ratio of Leu to Val in the LNAA supplement is greater  
4 than about 0.5:1.

1               11. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu and wherein the weight  
3 ratio of Leu to Val in the LNAA supplement is greater  
4 than 2:1.

1               12. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu; wherein the weight ratio  
3 of Leu to iLeu in the LNAA supplement is greater  
4 than about 0.5:1; and wherein the weight ratio of Leu to  
5 Val in the LNAA supplement is greater than about 0.5:1.

1               13. A method according to claim 12, wherein  
2 the weight ratio of Leu to iLeu in the LNAA supplement is  
3 greater than 3:1.

1           14. A method according to claim 12, wherein  
2 the weight ratio of Leu to Val in the LNAA supplement is  
3 greater than 2:1.

1           15. A method according to claim 6, wherein the  
2 LNAA supplement comprises Leu; wherein the weight ratio  
3 of Leu to iLeu in the LNAA supplement is greater than  
4 3:1; and wherein the weight ratio of Leu to Val in the  
5 LNAA supplement is greater than 2:1.

1           16. A method according to claim 6, wherein the  
2 LNAA supplement is substantially free from phenylalanine.

1           17. A LNAA supplement comprising Leu and Val  
2 in which the weight ratio of Leu to Val is greater than  
3 2:1.

1           18. A LNAA supplement according to claim 17,  
2 wherein the LNAA supplement is substantially free from  
3 phenylalanine.

1           19. A LNAA supplement comprising Leu and iLeu  
2 in which the weight ratio of Leu to iLeu is greater than  
3 3:1.

1           20. A LNAA supplement according to claim 19,  
2 wherein the weight ratio of Leu to Val in the LNAA  
3 supplement is greater than 2:1.

1           21. A LNAA supplement according to claim 20,  
2 wherein the LNAA supplement is substantially free from  
3 phenylalanine.

1                   22. A LNAA supplement comprising one or more  
2   LNAAs and further comprising Lys.

1                   23. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu.

1                   24. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu and wherein the  
3   weight ratio of Leu to iLeu in the LNAA supplement is  
4   greater than about 0.5:1.

1                   25. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu and wherein the  
3   weight ratio of Leu to iLeu in the LNAA supplement is  
4   greater than 3:1.

1                   26. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu and wherein the  
3   weight ratio of Leu to Val in the LNAA supplement is  
4   greater than about 0.5:1.

1                   27. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu and wherein the  
3   weight ratio of Leu to Val in the LNAA supplement is  
4   greater than 2:1.

1                   28. A LNAA supplement according to claim 22,  
2   wherein the LNAA supplement comprises Leu; wherein the  
3   weight ratio of Leu to iLeu in the LNAA supplement is  
4   greater than about 0.5:1; and wherein the weight ratio of  
5   Leu to Val in the LNAA supplement is greater than about  
6   0.5:1.

1                   29. A LNAA supplement according to claim 28,  
2 wherein the weight ratio of Leu to iLeu in the LNAA  
3 supplement is greater than 3:1.

1                   30. A LNAA supplement according to claim 28,  
2 wherein the weight ratio of Leu to Val in the LNAA  
3 supplement is greater than 2:1.

1                   31. A LNAA supplement according to claim 22,  
2 wherein the LNAA supplement comprises Leu; wherein the  
3 weight ratio of Leu to iLeu in the LNAA supplement is  
4 greater than 3:1; and wherein the weight ratio of Leu to  
5 Val in the LNAA supplement is greater than 2:1.

1                   32. A LNAA supplement according to claim 22,  
2 wherein the LNAA supplement is substantially free from  
3 phenylalanine.

1                   33. A LNAA supplement according to claim 22,  
2 wherein the LNAA supplement comprises, per 500 mg of LNAA  
3 supplement:  
4                   from about 100 mg to about 290 mg of Tyr;  
5                   from about 25 mg to about 75 mg of Trp;  
6                   from about 15 mg to about 50 mg of Met;  
7                   from about 15 mg to about 55 mg of iLeu;  
8                   from about 15 mg to about 50 mg of Threo;  
9                   from about 15 mg to about 55 mg of Val;  
10                  from about 15 mg to about 200 mg of Leu;  
11                  from about 10 mg to about 30 mg of His; and  
12                  from about 5 mg to about 200 mg of Lys.

1           34. A LNAA supplement according to claim 33,  
2 wherein the LNAA supplement comprises, per 500 mg of LNAA  
3 supplement, from about 10 mg to about 30 mg of Lys.

1           35. A LNAA supplement according to claim 33,  
2 wherein the LNAA supplement is substantially free from  
3 arginine.

1           36. A LNAA supplement according to claim 33,  
2 wherein the LNAA supplement is substantially free from  
3 phenylalanine.

1           37. A LNAA supplement comprising, per 600 mg  
2 of LNAA supplement:  
3           from about 100 mg to about 290 mg of Tyr;  
4           from about 30 mg to about 90 mg of Trp;  
5           from about 25 mg to about 75 mg of Met;  
6           from about 15 mg to about 45 mg of iLeu;  
7           from about 15 mg to about 50 mg of Threo;  
8           from about 15 mg to about 50 mg of Val;  
9           from about 40 mg to about 200 mg of Leu;  
10          from about 15 mg to about 45 mg of His; and  
11          from about 15 mg to about 50 mg of Arg.

1           38. A LNAA supplement according to claim 37,  
2 wherein the LNAA supplement further comprises Lys.

1           39. A LNAA supplement according to claim 37,  
2 wherein the LNAA supplement further comprises, per 600 mg  
3 of LNAA supplement, from about 5 mg to about 200 mg of  
4 Lys.

1                   40. A LNAA supplement according to claim 37,  
2 wherein the LNAA supplement is substantially free from  
3 phenylalanine.

1                   41. A method for treating a subject suffering  
2 from a condition involving a metabolic disorder involving  
3 the metabolism of a first amino acid X, said method  
4 comprising:  
5                   enterally administering to the subject a  
6 composition which is substantially free from said first  
7 amino acid X and which comprises a second amino acid Y  
8 that competes with amino acid X at a gastrointestinal  
9 tract transporter.

1                   42. A method according to claim 41, wherein  
2 the condition is not phenylketonuria and/or  
3 phenylalanemia.

1                   43. A method according to claim 42, wherein  
2 the gastrointestinal tract transporter is a Caco-2 cell  
3 transporter.

1                   44. A method according to claim 41, wherein  
2 the gastrointestinal tract transporter is a Caco-2 cell  
3 transporter.

1                   45. A method according to claim 44, wherein  
2 the condition is tyrosinemia; wherein the first amino  
3 acid X is tyrosine; and wherein the second amino acid Y  
4 is selected from Phe, Leu, Trp, Lys, His, and  
5 combinations thereof.

1           46. A method according to claim 44, wherein  
2 the condition is tyrosinemia; wherein the first amino  
3 acid X is selected from phenylalanine, tyrosine, and  
4 combinations thereof; and wherein the second amino acid Y  
5 is selected from Leu, Trp, Lys, His, and combinations  
6 thereof.

1           47. A method according to claim 44, wherein  
2 the condition is alkaptonuria; wherein the first amino  
3 acid X is selected from phenylalanine, tyrosine, and  
4 combinations thereof; and wherein the second amino acid Y  
5 is selected from Leu, Trp, Lys, His, and combinations  
6 thereof.

1           48. A method according to claim 44, wherein  
2 the condition is homocystinuria; wherein the first amino  
3 acid X is methionine; and wherein the second amino acid Y  
4 is an amino acid that competes with methionine at a  
5 gastrointestinal tract transporter.

1           49. A method according to claim 44, wherein  
2 the condition is a disorder affecting metabolism of a  
3 branched amino acid selected from leucine, isoleucine,  
4 valine, and combinations thereof; wherein the first amino  
5 acid X is selected from leucine, isoleucine, valine, and  
6 combinations thereof; and wherein the second amino acid Y  
7 is an amino acid that competes with the first amino acid  
8 X at a gastrointestinal tract transporter.

1           50. A method according to claim 49, wherein  
2 the condition is selected from maple syrup urine disease,  
3 isovaleric acidemia, methylmalonic acidemia, and  
4 propionic acidemia.



1                   51. A method according to claim 41, wherein  
2 said method further comprises:  
3                   restricting the subject's dietary intake of the  
4 first amino acid X.

1                   52. A method according to claim 41, wherein  
2 said method further comprises:  
3                   not restricting the subject's dietary intake of  
4 the first amino acid X.

1                   53. A method according to claim 52, wherein  
2 said enteral administration is carried out substantially  
3 at mealtime.

1                   54. A method according to claim 41, wherein  
2 said enteral administration is carried out substantially  
3 at mealtime.

1                   55. A method according to claim 41, wherein  
2 said enteral administration is carried out orally.